Johnson & Prudden

(W. B) (T. M)

MYXO-SARCOMA OF THE OPTIC NERVE WITH HYALIN DEGENERATION

BY

WALTER B. JOHNSON, M.D.

SURGEON TO THE PATERSON EYE AND EAR INFIRMARY

AND

T. MITCHELL PRUDDEN, M.D.

PATHOLOGIST TO THE MANHATTAN EYE AND EAR HOSPITAL

[Reprinted from the ARCHIVES OF OPHTHALMOLOGY, Vol. xiv., Nos. 2 and 3, 1885.]

MYXO-SARCOMA OF THE OPTIC NERVE WITH HYALIN DEGENERATION.

By WALTER B. JOHNSON, M.D.,

SURGEON TO THE PATERSON EYE AND EAR INFIRMARY.

AND

T. MITCHELL PRUDDEN, M.D.,

PATHOLOGIST TO THE MANHATTAN EYE AND EAR HOSPITAL.

(*With three illustrations.*)

IF we limit the designation "primary tumors of the optic nerve" to those tumors which originate within the optic-nerve sheath, there are recorded some thirty-seven cases, of which but a small number are reported in full. A large proportion of these, although appearing in the records with a varying nomenclature, seem to belong to the *sarcomata*, and a considerable number of them have shown a marked degree of *myxomatous* degeneration, or have been distinctly *myxo*-sarcomatous in structure.

The tumor which it is the purpose of this paper to record, though closely resembling others from the same situation fully described, presents some structural features worthy of special notice.

Carrie L. A., age twelve. First examined the patient, August 14, 1883. She is a strong, healthy, well-nourished child, with a good family history. Always enjoys good health and is accustomed to hard work. Presents herself with an *exophthalmos* of the left eyeball, which protrudes 13‴, at times pressing through the palpebral fissure and appearing to be entirely without the orbital cavity. The lower eyelid constantly everted is thickened

Reprinted from the ARCHIVES OF OPHTHALMOLOGY, Vol. xiv., Nos. 2 and 3, 1885.

and very much swollen, while coaptation of the lids is impossible. The conjunctiva is hypertrophied, red, and œdematous, and there is more or less continuous discharge of lachrymal secretion, containing shreds of thick *mucus*. The cornea is hazy, but not at all anæsthetic. The pupil moderately dilated responds very slightly to light. Dilatation is increased after instillation of atropine. The eyeball has considerable mobility in all directions, except downward. On palpation, a tumor is felt behind the globe, apparently occupying the entire cavity of the orbit, larger in the external and inferior quadrants. The eyeball is flattened from before backward, and is displaced upward, inward, and forward. The tumor is distinctly felt, gives the impression of being composed of numerous small bodies, about the size of peas, and has a soft, elastic feel. Pressure does not decrease the size of the tumor, and there is no pulsation or bruit.

The ophthalmoscopic examination shows a pink reflex ; retinal vessels very much diminished in size, and in the region of the macula, two or three small patches of choroidal atrophy, nearly as large as the head of a pin, direct method, with disk, very white and decreased in size.

The vision of R E = $\frac{20}{20}$. L E, no perception of light. The patient has a rapid pulse, about 120, but as no heart lesion is discovered, this is ascribed to excitement.

Previous History.—When a child six years old, her mother first noticed a prominence of the left eye, which has gradually increased until the present time. Six months after the first appearance of this enlargement, it was discovered that the eye was sightless.

The patient never had any pain or annoyance from her eye until one year ago (that is, five years after the trouble commenced), since which time she has been at times affected with a sense of fulness, and occasionally with pains about the eye and corresponding temple. These symptoms and more or less dizziness are aggravated on stooping. The patient was examined in consultation at the Manhattan Eye and Ear Hospital, and although there was some difference of opinion, the diagnosis, *orbital tumor of the optic nerve*, was made, and enucleation was advised, the majority believing the tumor malignant.

September 2, 1883.—The patient was anæsthetized and the operation commenced. The intention was to enucleate the tumor, without removing the eyeball, if possible. After the initiatory

steps had been performed, the very considerable size of the tumor compelled us to abandon the idea and proceed to enucleate the eyeball and tumor. The dissection of the conjunctiva was continued around the corneal margin. The fan-like attachments of the muscles exposed were 3‴ or 4‴ in breadth, and very much hypertrophied, and were easily cut without using a hook. The tumor, then exposed to view, behind a large quantity of orbital fat (which was probably the cause of the feeling of the small, pea-like bodies discovered on palpation), was solid, oblong, and almost entirely filled the orbit. For this reason it was difficult to make the dissection, and the external canthus was slit to make more room.

During the operation the nerve sheath, which was distended with fluid clear up to its junction with the globe, was punctured, and a yellowish serous fluid escaped.

The dissection was continued with the finger and the handle of a scalpel, until it was discovered that the tumor extended into the foramen of the optic nerve. Desiring to remove the entire tumor, a strong ligature was passed through it for the purpose of drawing it forward, so that, if possible, the section might be made behind it.

It seemed that the sheath, which enclosed the tumor, was attached to the optic foramen. An effort was made to cut it before severing the tumor, and then to drag the tumor forward and cut it. This was accomplished, as the surface of the tumor protruded nearly 1‴ from within the sheath after its excision. The result, however, was probably a recession of the arteries, as there was a hemorrhage after the operation, which at times would be considerable, and at other times only oozing, resisting for eight hours all measures adopted for its control. The patient vomited very frequently, and each effort at vomiting caused renewed bleeding, the vomited matter, for the most part, being blood, which had passed down through the floor of the orbit. Five hours after the hemorrhage had entirely ceased, and while the nurse was sleeping, there was a second hemorrhage, which almost resulted fatally. The examination of the orbit disclosed that its floor, thin and eroded from pressure, had broken through during the operation. The optic foramen seemed to be enlarged to such an extent that the point of the index finger could be introduced. It appeared free from tumor substance, although it was thought that some portion of the tumor may have been left behind. The wound healed kindly, but slowly, without much purulent discharge, and the tissues shrunk gradually back into the orbital cavity. For a num-

ber of weeks there remained an opening into the back of the orbit, which discharged a small quantity of thick *muco-pus*. Forty-eight days after the enucleation, this opening has healed. The patient is in good health. The orbital tissues present the usual amount of shrinkage, and the disease shows no tendency to recurrence.

May 1, 1885.—The patient is now nearly fourteen years old, and has enjoyed good health since the operation, September 2, 1883. She has not been troubled by any pain in the region of the operated eye, and has worn a glass eye for about a year.

The orbital tissues are slightly more shrunken than at the last examination, and there is no tumefaction in any part of the orbital cavity, nor any thing which would appear to indicate a probability of recurrence.

The tumor was examined microscopically by Dr. Prudden whose report follows.

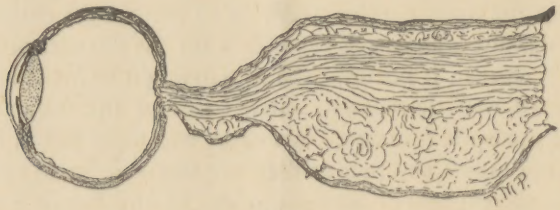

FIG. 1.

The tumor, after hardening in Müller's fluid and alcohol, presents the following macroscopical appearances:

It is irregularly ovoidal in shape, being somewhat flattened on one side. It is about 35 *mm.* long and about 20 *mm.* in the largest diameter. It is slightly nodular, and is completely invested by a dense, thick, connective-tissue sheath, which is continuous anteriorly with the sclerotic, and which is evidently the distended external optic-nerve sheath. This sheath is loosely attached to the tumor tissue within. From the anterior and more tapering end of the tumor, the optic nerve emerges, being somewhat narrower in diameter than normal and slightly flattened, and passing forward for about 1 *cm.* in a curved direction, enters the globe.

A longitudinal section through the middle of the tumor and the globe, presents the appearances seen in fig. 1.

Continuous with the optic nerve anteriorly, extending completely through the tumor, and lying somewhat to one side of its axis, is a longitudinally striated mass, about 8 *mm.* thick, somewhat denser than the remainder of the tumor, and for the most part sharply outlined against the rest of the tissue. This is evidently the enlarged optic nerve.

The tumor substance which surrounds the thickened optic nerve and occupies the space between it and the external sheath, is for the most part translucent, and appears to be made up of irregularly arranged masses and bands of soft tissue.

Microscopical examination of sections of this portion of the tumor surrounding the optic nerve, shows that it consists, in the main, of irregular larger and smaller masses and intricately interlacing bands of closely packed cells, separated by narrow and broad bands of dense fibrillar connective tissue, the whole moderately vascular. Into many of the cell-masses, connective-tissue fibres and fibrillæ penetrate ; delicate elastic fibres are also of frequent occurrence. In some places, notably immediately around the thickened optic nerve, the fibrillar connective tissue predominates, but in most parts the cell-masses make up most of the substance. In some parts of the tumor larger and smaller islets of distinctly myxomatous tissue are present, while in others the cell-masses appear to be simply œdematous, and thus present an appearance somewhat simulating that of mucous tissue.

Scattered everywhere among the cells of the tumor, in some places sparsely, in others in great numbers, are seen sections of very irregular, larger and smaller translucent, strongly refractile bodies with rounded contours, which appear to be in connection with the cells composing the above-described cell-masses, or with the smaller blood-vessels.

These translucent bodies have the general appearance of amyloid, but they are not stained in the manner characteristic of that substance by iodine or by methyl violet. They are, on the contray, very readily stained by eosin, fuchsin, —particularly the acid fuchsin,—methyl-green, and by hematoxylin, and retain these colors very tenaciously in the

presence of strong acetic acid, alcohol, and caustic potash (30 per cent.). When unstained they are but slowly affected by strong aqua ammoniæ and 33 per cent. caustic-potash solution. They are not very readily stained by carmine. Their general appearances and the above series of reactions would indicate that the strongly refractile bodies scattered through the tumor tissue belong to the material which results from the so-called hyalin degeneration, as described by Recklinghausen.[1]

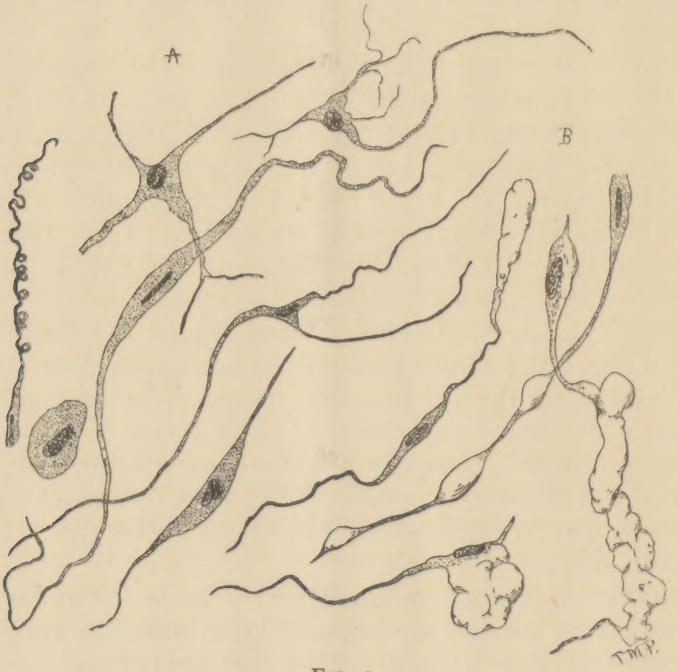

FIG. 2.

A. Cells from teased tumor. B. Degenerated cells from the same.

The shape of the cells composing the main part of the tumor, as well as the mode of origin of the above-described hyalin bodies, is best seen in bits of the tissue stained and carefully teased.

Some of the varied shapes of the cells prepared in this way are represented in fig. 2, A. The most abundant

[1] Recklinghausen in Billroth & Lücke's Deutsche Chirurgie, Lief. 2 and 3, page 404.

form is that of exceedingly long and slender spindles with either spheroidal or elongated nuclei. Some of these cells are so long and slender that they might readily be mistaken for simple fibres; careful scrutiny alone revealing the small nuclei. There are other fusiform cells which are broader and shorter, the bodies tapering to a point at a short distance from the nucleus. Branching cells with several processes, some long and fibre-like, others short and dividing and tapering, are quite numerous, as are also fusiform cells whose fibre-like ends are curled into close irregular spirals. Finally, larger and smaller spheroidal cells are present, but vary considerably in number in different parts of the tumor.

The teased preparations show, furthermore, in the most evident manner, the mode of origin of the hyalin material. This is formed, as shown in fig. 2, B., by an irregular degeneration of parts of the tumor cells. These are sometimes to a slight extent, sometimes almost completely, converted into the hyalin material, and are to a corresponding degree distorted and deformed. In some cases the whole thickness of the cell is not converted into the hyalin material, but the latter appears in the form of irregular shining masses or droplets completely enclosed by the cell protoplasm.

In the separated cells as well as in sections, the above-mentioned micro-chemical reactions were obtained.

Similar translucent masses have been described in tumors of the optic nerve by several observers. Structures which, judging from his drawings, were similar to these degenerated cells, were described by Perls,[1] as occurring in large numbers in a tumor of the optic nerve which he regarded as a true neuroma. The cells were considered to be ganglion cells; and the translucent shining material myelin in the process of formation about the fibre-like branches of the cells. The probability that this conception of the nature of the cells and translucent shining material by Perls was an error, has been pointed out by Vossius and Lebert,[2] who have described the same appearance in similar tumors of

[1] Perls: *Arch. für. Ophthal.*, Bd. xix., Abth 2, p. 287.

[2] Vossius: *ibid.*, Bd. xxviii., Abth. 3, p. 33.

the optic nerve, and who also had the opportunity of examining the original tumor of Perls. Lebert examined the translucent material in the fresh condition, and found that it did not stain black on treatment with osmic acid. If, as seems probable, this criticism is correct, a true neuroma of the optic nerve seems never to have been described.

Dr. Johnson's tumor was not received in the fresh condition, so that the usual micro-chemical reactions for fresh nerve tissue could not be employed; but a series of the tests applicable to preserved nerve tissue were made, all of which failed to give any indication of nerve elements in the tumor tissue proper, outside of the limits of the optic nerve. Among the tests above alluded to were the prolonged staining with dilute gold-chloride solution and reduction in caustic-soda solution; the use of the chloride of palladium and subsequent staining with carmine.

As the hyalin material retains the color imparted by acid fuchsin, it was not surprising that the acid fuchsin staining of Weigert should give a very excellent staining of the degenerated portions. It was, however, a surprise to find that the hematoxylin method of staining nerve tissue as described by Weigert gave most exquisite pictures of the topography of the degeneration; the hyalin material being stained bluish black, just as the myelin is, in successful preparations of the nerve tissue. This possibility of a similarity of reaction in hyalin material and in myelin must, therefore, evidently be borne in mind, if the possibility of error would be avoided in using either the hematoxylin or the acid fuchsin method of Weigert for staining nerve tissue.

The microscopical examination of the enlarged portion of the optic nerve, which passes through the tumor, shows that there is a considerable increase of the connective-tissue septa which divide the nerve into bundles, and also a very marked growth between the individual nerve fibres, apparently from the neuroglia, of cells similar to those composing the more characteristic parts of the tumor proper. The same hyalin degeneration is seen also in the new cells of the optic nerve, but it is most abundant in the periphery of

the bundles along the sides of the connective-tissue partitions of the nerve. The nerve fibres are in part normal, but, especially in the middle and posterior portions, they are to a considerable extent atrophied.

That part of the optic nerve which lies between the tumor and the globe shows simply a marked increase of the interstitial fibrillar connective-tissue, with corresponding atrophy of the nerve fibres, with here and there small masses of hyalin material, but none of the characteristic tumor cells.

In the tumor tissue proper there is hyalin degeneration of the walls or about the walls, especially of the smaller blood-vessels, as represented in fig. 3.

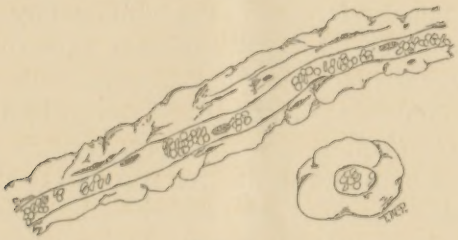

FIG. 3.

Capillary blood-vessels with hyalin degeneration of the walls.

Microscopical examination of the enucleated eye shows considerable increase of the connective tissue of the optic nerve, with corresponding atrophy of the nerve fibres and thickening of the walls of the retinal artery. There are patches of œdema of the retina, particularly of the outer layers. The choroidal blood-vessels are generally and deeply congested, and in the sections made there was found one small patch of atrophy of the choroid near the optic papilla. The eye appeared otherwise normal, but the nerve elements of the inner layers of the retina were not well preserved, so that their condition could not be determined.

The tumor apparently originated in the neuroglia of the optic nerve, or in the connective tissue of the inner sheath. Its growth was largely in the space between the external and internal nerve sheaths, and only to a moderate degree in the interstices of the nerve itself.

Recklinghausen reported hyalin degeneration of the blood-

vessels in one of the earlier cases. The translucent, shiny masses, as above stated, have been described and figured by several observers. Vossius conjectures that they might be of a "hyalin-colloid" nature; but otherwise the nature of the material has been but indefinitely characterized.

It will thus be seen, if the interpretation placed by the writer upon the character of the degenerated portions of these tumors be correct, that hyalin degeneration, which is of occasional occurrence in a great variety of tumors, is quite frequent in the myxo-sarcomata of the optic nerve, and may give rise, here as elsewhere, to very peculiar and oftentimes puzzling microscopical appearances.

It is not necessary for the purposes of the present paper to make a detailed reference to the bibliography of tumors of the optic nerve. Twenty-seven cases were collected by Willemer, *Archiv für Ophthal.*, Bd. xxv., Abth. 1, p. 161, 1879. This number was increased to thirty-six by Vossius, *Archiv für Ophthal.*, Bd. xxviii., 1882, and in his article references may be found to the literature up to that date.

Another case has since been described by Veron, *Recueil d'Ophthalm.*, Jan., 1883, Reviewed in ARCHIVES OF OPHTHALMOLOGY, vol. xiii., No. 2, p. 283.

G. P. PUTNAM'S SONS, PRINTERS
NEW YORK